

SHORT REPORT

Status-like recurrent pilomotor seizures: case report and review of the literature

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Abstract

A diabetic 66 year old man who presented with pilomotor seizures in his right hemibody is described. The seizures recurred with an increasing frequency, leading to a status-like condition associated with Korsakoff's syndrome. An EEG was performed and several electroclinical seizures were recorded. Brain MRI was negative. The patient, who was treated with carbamazepine, became seizure free after 1 week. Memory and behaviour gradually returned to normal within 3 weeks. There was no further neurological episode during an 8 year follow up. Hyperosmolar, non-ketotic hyperglycaemia was considered to be the cause of the seizures. The pathophysiology of pilomotor seizures is discussed and the literature on the subject reviewed.

(J Neurol Neurosurg Psychiatry 2000;68:647-649)

Keywords: pilomotor seizures; autonomic epilepsy; Korsakoff's syndrome; hyperosmolar non-ketotic hyperglycaemia

Horripilation, piloerection, cutis anserina, and gooseflesh are among the many names used to designate erection of body hairs in animals or humans. This skin response to fear, cold, rage, or sexual excitement can also be an epileptic symptom, constituting, in its purest form, a pilomotor seizure. Since Féré's initial description in 1896,¹ only 20 cases of this type of autonomic seizure have been reported. We report a new case of particular clinical interest, because it seems to be the first case attributable to hyperosmolar, non-ketotic hyperglycaemia.

Case report

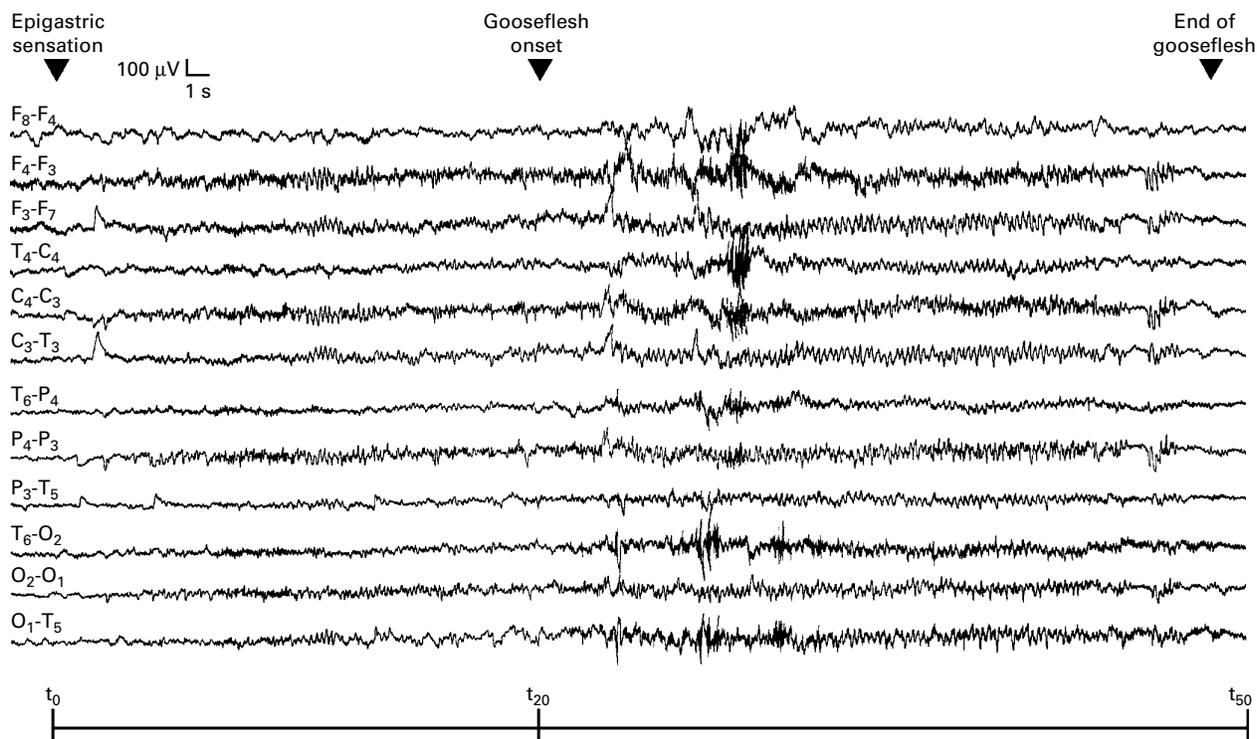
A 66 year old right handed retired policeman was admitted to an internal medicine ward for "unexplained bouts of chills" occurring several times a day. Investigations did not detect an infectious disease. Although temperature, erythrocyte sedimentation rate, and white cell count were normal and blood cultures remained negative, intermittent bacteraemia was considered as a possible diagnosis. Antibiotic therapy was given for 2 weeks with no effect on the chill sensation. Three weeks later, after an emotional shock (he had just learned of his daughter's impending divorce), the frequency of chills increased and behavioural disturbances were

noted. The patient was given 30 IU insulin per day for insulin requiring diabetes mellitus. He consumed 50 g alcohol daily. On examination in the neurology department, there were 20 to 30 spells of chill an hour, each lasting from 10 seconds to 2 minutes. During a spell, several events occurred in the following sequence: (a) a peculiar epigastric sensation with a feeling of thoracic oppression; (b) flushing of face and arms; (c) a sensation of hot liquid in the right hand, spreading to the right foot and then to the whole body; (d) gooseflesh over the right hemibody. Spells were often triggered by an emotional factor. During the spells, the patient remained conscious and able to execute commands. There was neither diaphoresis, nor a change in blood pressure or cardiac rate. When examined between two spells, the patient was agitated and disorientated as to time and place. There were false recognitions, confabulations, and delusion of persecution. He could not memorise new information (for example, he was unable to remember what he had just eaten). By contrast, the patient could state his age correctly and had no difficulty in recalling various facts learned long before (such as the dates of the two world wars and even the date of the battle of Waterloo!). Verbal expression and oral comprehension were normal, as were writing and drawing. Digit span, verbal fluency (16 names of animals in one minute) and "abstract thinking", tested by proverb interpretation and similarities, were within normal limits. There was neither apraxia nor agnosia. Neurological and general examination were otherwise unremarkable. Therefore, autonomic seizures were diagnosed. An EEG recorded several electroclinical seizures. During a seizure, sharp wave discharges at theta frequencies started a few seconds before the onset of gooseflesh and were usually recorded in the left frontotemporal region before spreading throughout both hemispheres. They ended abruptly without slow postictal alteration (figure). Routine laboratory examination showed hyperglycaemia (10.2 mmol/l) and glucosuria without ketosuria. Blood cell count was normal, as were erythrocyte sedimentation rate (11 mm/h) and blood electrolytes. Osmolarity was 317 mmol/l (normal <290 mmol/l). Alkaline phosphatase, alanine aminotransferase, aspartate aminotransferase, and bilirubin were within normal limits. There was a slightly raised γ -glutamyl transferase concentration (46 U; normal <38 U). Syphilis and HIV serologies

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Received 26 August 1999
and in revised form
15 December 1999
Accepted 5 January 2000



EEG recording of a pilomotor seizure lasting 1 minute. Note the appearance, about 10 seconds after the initial symptom (epigastric sensation), of sharp wave discharges in the 4–6/s range, starting in the left frontotemporal region. The gooseflesh phenomenon occurred 9 seconds later.

were negative. Brain CT and MRI were normal. Examination of CSF disclosed hyperproteinorhachia (0.70 mmol/l) with normal cellularity (1/mm³) and normal glycorrachia (6.1 mmol/l). Despite insulin therapy, glycaemia remained unstable and above the normal range (7 to 18 mmol/l), but osmolarity returned to a level below 300 mmol/l within 4 days. The patient was treated with carbamazepine (800 then 1200 mg per day) in addition to intravenous vitamins (B1, B6, nicotinamide). The seizures became less frequent and disappeared within a week. Behaviour returned to normal. One month later,

some mild memory difficulties were still present and thereafter remained unchanged. Eight years later the patient died of cardiac failure. There was no postmortem examination.

Discussion

Among the different subtypes of autonomic seizures, pilomotor seizure is one of the least common, only 20 cases having previously been reported (table).^{1–17} Its incidence is probably underestimated, as it may, as in the present case, simulate an infectious disorder. Pilomotor seizures are commonly accompanied by other

Table 1 Literature review of test results

Case report (first author/year of publication)	Age/sex	Aetiology or associated neurological disease	EEG findings	Associated symptoms*
Féré ¹ (1896)	?/M	Seizures since infancy	—	sud/the/los/sei
Hutchinson ¹¹ (1898)	50/M	Neurosyphilis	—	the/chi/sei
Féré ¹⁰ (1904)	53/M	Ophthalmoplegic migraine	—	sen/sei/sud/hea
Penfield ¹² (1929)	41/F	Pearly tumour of the third ventricle	—	chi/sud/crs/los
Landau ¹³ (1953)	29/M	CNS involvement by streptococcal septicaemia	Theta bursts (interictal)	the/nps
Mulder ⁵ (1954)	43/M	Grand mal	L temporal slow waves	Not described
Mulder ⁵ (1954)	25/F	Seizures since infancy	L temporal sharp waves (after metrazol)	crs/chi/los
Penfield ¹⁴ (1954)	?/F	Unknown	—	gis/sud/crs
Brody ⁴ (1960)	29/F	Astrocytoma of L temporal lobe	—	crs/sei
Brody ⁴ (1960)	53/F	Glioblastoma of R parieto-occipital lobe, thalamus and basal ganglia	R temporal theta rhythmic activity (ictal and interictal)	gis/the/los/sen
Andermann ¹⁵ (1984)	61/M	Glioblastoma of R temporal, frontal and insula lobe	—	nps/crs/tri
Green ⁶ (1984)	44/M	Glioblastoma of R temporal lobe	R frontotemporal 5 Hz spikes and waves (ictal)	the/chi/gis/sen
Lesser ¹⁶ (1985)	35/M	L temporal lobectomy	L temporal paroxysmal alpha activity (ictal)	sei/chi/sen/tri
Tyndel ¹⁰ (1986)	56/M	Glioblastoma of L parieto-temporal lobe	—	chi
Brogna ² (1986)	58/M	Astrocytoma of L mediobasal temporal lobe	L temporal rhythmic sharp wave (ictal, NPE)	hea/sud/gis/sen
Ahern ⁸ (1986)	35/M	R anterior temporal lobe post-traumatic contusion	Spike discharges (interictal, NPE)	the/nps
Dupuy ⁷ (1989)	56/M	Bilateral deep temporal radionecrosis	Generalised theta rhythmic activity (ictal)	sen/nps/tri/gis
Scopetta ³ (1989)	58/M	Glioma of L deep temporal lobe	L temporal theta rhythmic activity (ictal)	hea/sud/nps/gis
Yu ¹⁷ (1998)	29/M	Hippocampal sclerosis	L temporal spikes	chi
Yu ¹⁷ (1998)	33/M	Meningioma of the sphenoid region	L temporal rhythmic discharges	the/nps
Present case	66/M	Non-ketotic hyperglycaemia ?	Generalised theta rhythmic activity (ictal)	nps/sei
				los/nps/crs/chi
				chi/the/tri
				nps/gis/crs

*crs=Cardiorespiratory symptoms; chi=chills; gis=gastrointestinal symptoms; hea=cephalic sensation (head); los=loss of consciousness; tri=emotional triggering factor; nps=neuropsychological disorders; sei=other kinds of seizure; sen=sensory symptoms; sud=sudomotor activity; the=thermal sensation; NPE=nasopharyngeal electrode.

thermoregulatory phenomena (chills, sudomotor activity, thermal sensation) and by a peculiar epigastric sensation. No specific EEG pattern has been reported, but ictal theta discharges, as seen in our patient, are common.^{3 4 6 7} Carbamazepine or phenytoin was effective in all but two cases.^{6 17} Of these 20 previously reported cases, a clear cause was found in 13, including gliomas (n=7), pearly tumour (n=1), meningioma (n=1), lobectomy for intractable seizures (n=1), post-traumatic cerebral contusion (n=1), radionecrosis (n=1), and hippocampal sclerosis (n=1). In six cases, a previous neurological history was mentioned: long standing seizure disorder (n=3), neurosyphilis (n=1), ophthalmoplegic migraine (n=1), and cerebral damage after streptococcal septicaemia (n=1). In one case, no details of presumed aetiology or medical history were given. The aetiology of epilepsy in the present case poses a problem. Cerebral MRI was normal and an infratentorial tumour could be excluded as no further neurological episode was seen during an 8 year follow up. Chronic alcoholism, although moderate, may have played a part as this condition can cause both partial and generalised seizures. Hyperosmolar, non-ketotic hyperglycaemia can induce pharmacoresistant focal seizures which are usually controlled by normalising osmolarity and glycaemia.¹⁸ In the present case, seizures disappeared while osmolarity was going down, although glycaemia remained above normal, and the time course of recovery (over a week) was typical of the seizures related to hyperosmolar, non-ketotic hyperglycaemia. Furthermore, non-recurring episodes in long term follow up suggest an acute, provoked cause of partial status epilepticus such as hyperosmolar, non-ketotic hyperglycaemia. As far as the role of carbamazepine is concerned, it may have been coincidental. We finally concluded that hyperosmolar, non-ketotic hyperglycaemia was the most probable cause of our patient's seizures.

The pathophysiology of pilomotor seizures is still a subject of debate. The existence of pilomotor seizures and the ability of a few normal people to voluntarily produce or inhibit erection of the body hairs,^{19 20} suggest that pilomotor activity is under the control of the cerebral cortex. However, it is not known precisely which structures are involved in the regulation of this phenomenon. From the results of animal experiments and observations in humans, several structures seem to be involved: the hypothalamus, the limbic system and the frontal lobe, in particular the orbital cortex and the premotor area.^{19 21-25}

Hypothalamus—Piloerection can be produced by electrical or pharmacological stimulation of the hypothalamus or abolished by bilateral hypothalamectomy in cats.^{21 22}

Limbic system—Piloerection was elicited by electrical stimulation of the amygdaloid nuclei in unanaesthetised cats and by excitation of the cingulate gyrus in monkeys.^{23 24}

Orbital cortex—As it is involved in other autonomic responses, the orbital cortex is likely to be a control centre for piloerection, although the

phenomenon has not been studied and described in experimental stimulation of this zone.²⁵

Premotor area—Lindsley and Sassaman recorded changes in brain potentials in the premotor area during voluntary piloerection in a human and Messimy noted an exaggerated pilomotor reaction after removal of the premotor area in monkeys.^{19 22}

The transient impairment of anterograde memory presented by our patient is an interesting corollary to this report. The fact that other intellectual capacities were preserved indicates that the anterograde amnesia did not form part of a postictal confusional state, but was a selective memory disorder consistent with the definition of Korsakoff's syndrome. Although high alcohol consumption may have contributed to Korsakoff's syndrome, his intake was not grossly excessive and a good recovery of the memory disturbance is unusual for that condition. Memory disturbances have been described in association with pilomotor seizures,^{2 4 9} and we would favour the argument that an ictal or postictal phenomenon was of more relevance in his case.

We thank Dörte Clausen for her assistance in the preparation of this manuscript.

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