SHORT REPORT

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

Emmanuel Roze, Paul Oubary, François Chédru

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.

Service de Neurologie, Centre Hospitalier Général de Meaux, 6-8 rue Saint-Fiacre, BP 218, 77108 MEAUX Cedex, France
E Roze
P Oubary
F Chédru

Correspondence to:
Dr F Chédru
f-chedru@ch-meaux.fr

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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 phenomenon. It can constitute, 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 disoriented 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
were negative. Brain CT and MRI were normal. Examination of CSF disclosed hyperproteinorhachia (0.70 mmol/l) with normal cellularity (1/mm³) and normal glycorrhachia (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

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**Table 1 Literature review of test results**

<table>
<thead>
<tr>
<th>Case report (first author/year of publication)</th>
<th>Age/sex</th>
<th>Aetiology or associated neurological disease</th>
<th>EEG findings</th>
<th>Associated symptoms*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Féré1 (1896)</td>
<td>5/M</td>
<td>Seizures since infancy</td>
<td>—</td>
<td>sud/the/los/hei</td>
</tr>
<tr>
<td>Hutchinson1 (1898)</td>
<td>50/M</td>
<td>Neurophilis</td>
<td>—</td>
<td>the/chi/</td>
</tr>
<tr>
<td>Féré2 (1904)</td>
<td>53/M</td>
<td>Ophthalmoplegic migraine</td>
<td>—</td>
<td>sen/los/hea</td>
</tr>
<tr>
<td>Penfield3 (1929)</td>
<td>41/F</td>
<td>Pearly tumour of the third ventricle</td>
<td>Theta bursts (interictal)</td>
<td>chi/los</td>
</tr>
<tr>
<td>Landau4 (1933)</td>
<td>29/M</td>
<td>CNS involvement by streptococcal septicaemia</td>
<td>L temporal slow waves</td>
<td>the/np</td>
</tr>
<tr>
<td>Mulder5 (1954)</td>
<td>43/M</td>
<td>Grad mal</td>
<td>Not described</td>
<td></td>
</tr>
<tr>
<td>Mulder6 (1954)</td>
<td>25/F</td>
<td>Seizures since infancy</td>
<td>L temporal sharp waves (after metrazol)</td>
<td>cri/chi/los</td>
</tr>
<tr>
<td>Penfield7 (1954)</td>
<td>35/F</td>
<td>Unknown</td>
<td>—</td>
<td>cri/</td>
</tr>
<tr>
<td>Brody8 (1960)</td>
<td>29/F</td>
<td>Astrocytoma of L temporal lobe</td>
<td>L temporal theta rhythmic activity (ictal and interictal)</td>
<td>sei/gis/nps/crs/cri</td>
</tr>
<tr>
<td>Andermann9 (1984)</td>
<td>61/M</td>
<td>Glioblastoma of R temporal, frontal and insula lobe</td>
<td>—</td>
<td>the/chi/</td>
</tr>
<tr>
<td>Green10 (1984)</td>
<td>44/M</td>
<td>Glioblastoma of R temporal lobe</td>
<td>L frontotemporal 5 Hz spikes and waves (ictal)</td>
<td>sei/cri/</td>
</tr>
<tr>
<td>Lesser11 (1985)</td>
<td>35/M</td>
<td>L temporal lobectomy</td>
<td>L temporal paroxysmal alpha activity (ictal)</td>
<td>sei/cri/</td>
</tr>
<tr>
<td>Tyndall12 (1986)</td>
<td>56/M</td>
<td>Glioblastoma of L parieto-temporal lobe</td>
<td>—</td>
<td>hea/</td>
</tr>
<tr>
<td>Brogna13 (1986)</td>
<td>58/M</td>
<td>Astrocytoma of L mediobasal temporal lobe</td>
<td>L temporal theta rhythmic activity (ictal), NPE</td>
<td>hea/</td>
</tr>
<tr>
<td>Abern14 (1986)</td>
<td>35/M</td>
<td>R anterior temporal lobe post-traumatic contusion</td>
<td>L temporal paroxysmal sharp wave (ictal, NPE)</td>
<td>nps/cri/</td>
</tr>
<tr>
<td>Dupuy15 (1989)</td>
<td>56/M</td>
<td>Bilateral deep temporal radionecrosis</td>
<td>Generalised theta rhythmic activity (ictal)</td>
<td>hea/np</td>
</tr>
<tr>
<td>Scopetta16 (1989)</td>
<td>58/M</td>
<td>Glioma of L deep temporal lobe</td>
<td>L temporal paroxysmal sharp (ictal, NPE)</td>
<td>nps/cri/</td>
</tr>
<tr>
<td>Yu17 (1998)</td>
<td>29/M</td>
<td>Hippocampal sclerosis</td>
<td>L temporal rhythmic spikes</td>
<td>nps/cri/</td>
</tr>
<tr>
<td>Yu18 (1998)</td>
<td>33/M</td>
<td>Meningioma of the sphenoid region</td>
<td>L temporal rhythmic spikes</td>
<td>nps/cri/</td>
</tr>
<tr>
<td>Present case</td>
<td>66/M</td>
<td>Non-ketotic hyperglycaemia</td>
<td>Generalised theta rhythmic activity (ictal)</td>
<td>nps/cri/</td>
</tr>
</tbody>
</table>

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*crs=Cardiorespiratory symptoms; chi=chills; gis=gastrointestinal symptoms; hei=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. Carbamazepine or phenytoin was effective in all but two cases. 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), neurosphilis (n=1), ophthalmomoplegic migraine (n=1), and cerebral damage after streptococcal septicemia (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 infraimaging phenomenon has not been studied and described in experimental stimulation of this zone.

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. 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 Korsako’s syndrome. Although high alcohol consumption may have contributed to Korsako’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, and we would favour the argument that an ictal or postictal phenomenon was of more relevance in his case.

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