Semiology of the Rare Seizure Subtype Piloerection

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Objective: To report piloerection and vasomotor instability as a sole manifestation of partial seizures.

Design: Case report.

Setting: Inpatient tertiary care center.

Patient: A 72-year-old man with acute onset of repetitive autonomic events.

Main Outcome Measures: Extensive cardiovascular evaluation, electroencephalographic testing, radiology, laboratory assessment, and frequency of clinical events (seizures).

Results: We characterize an uncommon manifestation of right temporal lobe partial seizures, initially thought to be cardiovascular in origin, as pilomotor seizures accompanied by other autonomic phenomena. The ictal electroencephalogram recordings established the diagnosis of right temporal lobe seizures, and head magnetic resonance imaging demonstrated right mesial temporal T2 signal change, enhancement, and subsequent atrophy. The underlying etiology was not uncovered despite an extensive laboratory and radiological evaluation. However, given the history and imaging findings, an infectious or secondary immunological etiology was suspected.

Conclusions: Pilomotor events with other transient autonomic features, such as tachycardia and blood pressure fluctuations, may represent localization-related epilepsy. The acute onset of these events accompanied by other autonomic phenomena in the setting of focal magnetic resonance imaging abnormalities within the hippocampal region raises questions concerning pilomotor seizures as well as the differential diagnosis of acute seizure activity in the setting of neuroimaging findings characteristic for limbic encephalitis.

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A utonomic fluctuation is a relatively common phenomenon in seizures. Tachycardia, bradycardia, blood pressure changes, gastrointestinal disturbance, and respiratory fluctuations are relatively familiar occurrences during seizures.1-4 “Goose bumps,” however, are a less recognized autonomic manifestation of seizure (pilomotor seizure).4-8 Moreover, when present, autonomic features usually accompany more clearly recognized ictal features (loss of awareness, focal motor activity), making an epilepsy diagnosis difficult when autonomic symptoms are the sole presenting feature.

REPORT OF A CASE

A 72-year-old right-handed man with a medical history of hyperlipidemia and gastroesophageal reflux disease presented to our emergency department 1 week after acutely developing escalating spells of tachycardia, hypertension, warmth, facial flushing, and right hemibody piloerection. Accompanied with these sensations was a feeling of palpitations, anxiety, and occasional headache. By the time of presentation, his spells were occurring up to 25 to 30 times per day and lasting roughly 30 seconds. He denied any alteration of consciousness, any involuntary motor activity, focal weakness, or other neurologic or systemic concerns. With these episodes, he was noted to have severe fluctuations in blood pressure and heart rate, with systolic blood pressure as high as 210 mm Hg and heart rate as high as 120 beats/min (baseline systolic blood pressure was typically between 120-150 mm Hg and heart rate was around 80 beats/min).

Because of the possibility of a cardiac or endocrine etiology of his symptoms, the patient was admitted to the inpatient cardiology service for further evaluation and monitoring. Initial laboratory studies revealed a normal complete blood cell count,
electrolyte study results, glucose level, cardiac biomarkers, thyroid and liver function test results, C-reactive protein level, and erythrocyte sedimentation rate. Electrocardiogram showed a normal sinus rhythm at baseline. Twenty-four–hour urine catecholamine analysis results and fractionated metanephrine and vanillylmandelic acid levels were also obtained and found to be normal. Computerized tomography of the chest, abdomen, and pelvis was unrevealing.

Because his cardiac and endocrine evaluation was unremarkable, a neurology consultation was obtained to determine if these spells could be neurologic in origin. The patient had no risk factors for epilepsy, such as a personal or family history of seizures, febrile seizures, head trauma, or history of meningitis, encephalitis, or stroke. Initial neurologic examination, including formal mental status and language evaluation, was entirely normal. No focal or lateralizing findings were noted on neurologic examination. Examination during a typical spell again demonstrated normal mental status, intact language, and no focal or lateralizing findings. However, marked piloerection in the right upper and lower extremity, with milder findings in the left hemibody, was seen.

An electroencephalogram was obtained on the second hospital day because of the possibility that these spells represented seizures, despite no other clear seizure manifestations. During monitoring, 5 stereotyped electrographic seizures were recorded that correlated with the patient’s typical spells. During each event, the electroencephalogram showed right temporal sharp waves that evolved into high frequency discharges that then spread to the entire right hemisphere. The seizure discharge would then spread to involve the contralateral temporal lobe (Figure 1). No cognitive impairment was noted during any of the 5 electrographic events. Given these findings, the patient was treated with intravenous lorazepam and fosphenytoin and eventually switched to oral levetiracetam monotherapy, resulting in complete cessation of his seizures.

Magnetic resonance imaging of the brain performed 3 days after hospital presentation showed T2 signal abnormality with faint gadolinium enhancement of an enlarged right hippocampal formation (Figure 2). Cerebrospinal fluid analysis demonstrated cerebrospinal fluid protein elevation to 68 mg/dL (range, 14–45 mg/dL), but normal total nucleated cell counts, glucose level, gram stain results, IgG index, and IgG synthesis rate and no unique cerebrospinal fluid oligoclonal bands. Cerebrospinal fluid viral studies for herpes simplex, cytomegalovirus, Epstein-Barr virus, and varicella zoster were negative. Further serologic evaluation revealed negative syphilis, Lyme, and serum angiotensin converting enzyme test results. Results of a serum paraneoplastic panel that included voltage-gated potassium channel antibody level were normal. Whole-body fludeoxyglucose F 18–positron emission tomography scan was performed

Figure 1. Scalp electroencephalogram tracing displayed in longitudinal bipolar montage. The electroencephalogram demonstrates a right temporal onset (T8) rhythmic discharge that spreads to involve the central midline (Cz) and midline frontal (Fz) electrodes. The onset of the seizure was clinically associated with piloerection. bpm indicates beats per minute; HR, heart rate.
to assess for occult tumor and demonstrated no obvious site of malignancy.

The patient initially did well; unfortunately, 3 months after initial evaluation, he presented again with frequent, short, intermittent spells of piloerection and autonomic fluctuation. A second electroencephalogram confirmed the events were again partial seizures. Head magnetic resonance imaging now showed atrophy of the right hippocampus and resolution of the previously seen gadolinium enhancement but continued faint T2 signal abnormality involving the right hippocampal formation (Figure 3). Despite maximization of his levetiracetam dose, he did not have resolution of seizures. Valproic acid was added as a second agent, resulting in termination of his seizures, and eventually was discontinued. The patient remains seizure free with levetiracetam monotherapy. The etiology of his acute-onset simple partial seizure disorder and magnetic resonance imaging abnormalities remains in question but is presumed to be secondary to a resolving infectious or inflammatory process.

**COMMENT**

Autonomic manifestations of seizures are not uncommon. Symptoms of heart rate changes, blood pressure fluctuation, palpitations, anxiety, and gastrointestinal dysfunction with seizures have been reported but are frequently accompanied by other motor and cognitive signs that raise the possibility of seizures as an etiology. The patient's headache during the event was likely related to hypertensive urgency due to vasomotor changes secondary to the seizure. Our case is interesting in that the autonomic symptoms were the only ictal presentation and highlight the diagnostic challenge in such cases.

The central autonomic network has yet to be clearly defined, but limbic areas, including the insula, cingu-
late gyrus, hippocampus, amygdala, orbitofrontal cortex, and connections with the hypothalamus and brainstem autonomic centers, appear to play a role in these seizure types.\textsuperscript{1,4-7,9} Therefore, epileptic discharges from these areas may be responsible for autonomic symptom sequela. A relatively uncommon presentation of autonomic seizures appears to be piloerection, and previous reports have suggested that piloerection may be a rare ictal manifestation in patients with temporal lobe epilepsy.\textsuperscript{5-8} A prior retrospective study of pilomotor seizure suggested that unilateral piloerection was frequently seen ipsilateral to the epileptic focus, which can be of localizing value in presurgical planning.\textsuperscript{9} Our patient had acute and recurrent right hemibody pilomotor seizures and right hippocampal signal change with enhancement, consistent with this theory. The spread of the piloerection to the contralateral hemibody in our patient was most likely due to spread of epileptic discharge through the autonomic network.

Diagnostic and treatment implication in new-onset seizures in adults, particularly elderly individuals, can be challenging. The prevalence of epilepsy progressively increases after the age of 60 years and several risk factors and etiologies account for this, including stroke, tumors, and neurodegenerative disease, as well as increased incidence of inflammatory, infectious, and possibly autoimmune conditions.\textsuperscript{10-12} In the case presented, we see evidence of presumed acute mesial temporal signal change and enhancement followed several months later by resolution of enhancement and subsequent progressive atrophy.\textsuperscript{13} Given the location and imaging characteristics, the term \textit{limbic encephalitis} might be applied. The diagnosis of this entity is based on clinical, neuropathologic, and neuroimaging characteristics. Typical clinical and electrophysiological features in limbic encephalitis can include memory impairment, personality changes, psychiatric symptoms, and temporal lobe seizures. Head magnetic resonance imaging often shows findings of unilateral or bilateral swollen limbic structures demonstrating signal change followed several months to years later by atrophy similar to our case.\textsuperscript{14} If concern for limbic encephalitis is present, evaluation for infectious, paraneoplastic, and nonparaneoplastic inflammatory conditions should be undertaken.\textsuperscript{12-14} Unfortunately, despite extensive testing in our patient, no specific etiology for these new-onset seizures could be identified; however, given the imaging characteristics, and acuity of presentation, frequent monitoring for underlying neoplastic and autoimmune conditions in our patient will continue.

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