Piloerection: A Rare Ictal Phenomenon – Case Report and Review of Literature

Sir,

We report the case of a 28-year-old man who presented with spells of unusual nature for 8 months. These events were characterized by frequent brief episodes of fearfulness and sensation of panic associated with goose bumps or piloerection over the left upper limb with unusual cold sensation going down his spine, palpitations, brief breathing difficulty and brief change in voice. He was completely aware and communicative during the episodes which lacked other autonomic or motor symptoms, lasted for 30 s-1 min and occurred 10–20 times a day. Although mainly spontaneous, there was increased frequency during periods of mental stress. An additional history of loss of consciousness with probable tonic-clonic limb movements was elicited at the very onset of this illness. However, no specific antiepileptic drug had been given. There was no febrile seizure, head trauma, encephalitis, meningitis, or cerebrovascular disease in the past. Family history was not contributory. The patient had attributed these symptoms to certain significant recent stressful life events and his inability to deal them. He initially consulted a psychiatrist and was treated as a case of panic disorder with anxiolytics and beta-blockers with marginal improvement. After 8 months, one of the authors considered the possibility of partial seizures with autonomic features though psychogenic nonepileptic events could not be ruled out completely. Long-term video-electroencephalography (EEG) monitoring showed intermittent theta (5–6 Hz) activity and frequent spike wave discharges over the left frontotemporal region. Ten habitual events characterized by piloerection over his left forearm with the sensation of cold running down his spine were recorded while conscious and responsive. No orofacial or limb automatism, speech arrest or behavioral arrest was noted. Ictal EEG showed ictal onset over left frontotemporal region preceding the clinical event by 10–15 s [Figure 1a and b]. Magnetic resonance imaging brain showed features suggesting left mesial temporal sclerosis. The patient was initiated on antiepileptic drugs (lamotrigine and clobazam with gradual dose escalation) and behavioral counseling with 80% seizure

Table 1: Literature review of 11 patients with ictal piloerection and delay in diagnosis

Serial number	Author	Age	Gender	Only thermoregulation involved	Other aura	Other sz type	Epilepsy duration	Frequency	Etiology	Follow-up since diagnosis	Outcome	Misdiagnosis/ delayed diagnosis
1	Roze *et al*.[3]	66	Male	No	Epigastric	No	Few days	20-30/h	Nonketotic hyperglycemia	8 years	Sz free on AEDs	Yes, intermittent bacteremia
2	Cutts *et al*.[5]	49	Male	No	Gustatory, epigastric	None	1 year	8-12/day	Astrocytoma	15 months	Sz free on AEDs, postsurgery	Yes, as depression
3	Dove *et al*.[7]	26	Female	Yes	No	CPS	15 years	3-4/h	Mesial temporal sclerosis	NA	Sz controlled on AEDs	Yes, as migraine
4	Puligheddu *et al*.[8]	54	Male	Yes	No	None	3 years	1/week	Idiopathic	3 years	Sz controlled on AEDs	Probably neurovegetative dysfunction, patient on BZDs for 3 years before
5	Mittal *et al*.[9]	57	Male	Yes	Occasional olfactory	None	2 years	15/day	Grade 2 astrocytoma with oligodendroglial component	2 years	Sz free postsurgery	Yes delayed diagnosis for 2 years, sought late medical attention
6	Lam *et al*.[10]	72	Male	Yes	Tachycardia	None	1 week	25-30/day	Probable nonparaneoplastic limbic encephalitis Nonparaneoplastic limbic encephalitis	NA	Sz free on AEDs	Yes, as cardiovascular event
7	Haykal and Abou-Khalil[11]	75	Male	Yes	Remote memories recall, dysarthria	None	2 months	10/day	Postherpes zoster encephalitis	3 month	Sz free on AEDs	Yes, as nonepileptic
8	Kurita *et al*.[12]	38	Male	Yes	No	None	1 month	10/day	Infection/ inflammation	1 years	Sz free on AEDs	Yes, as autonomic dysfunction
9	Asha *et al*.[13]	66	Male	No	Visual, detachment from reality, olfactory and gustatory		1 month	4-5/3 weeks	Intraventricular glioblastoma multiforme	14 months	Sz free postsurgery till death	Yes, ? Due to eating prawns
10	Rocamora *et al*[6]	52	Male	Yes	No	None	4 years	1-3/day	Anti-LGi 1 limbic encephalitis	2.4 years	Refractory seizures despite steroids, IVIg	Yes, as psychiatric disorder
11	Panda (present case)	28	Male	Yes	No	None	8 months	10-20/day	Mesial temporal sclerosis	2 years	Frequency decreased initially on AEDs, again refractory	Yes, as panic disorder

CPS = Complex partial seizure, NA = Not available, sz = Seizure, AEDs = Antiepileptic drugs, IVIg = Intravenous immunoglobulin, BZDs = Benzodiazepines, Anti-LGi 1= Anti-Leucine-rich Glioma inactivated-1

reduction at 6 months follow-up. The patient's family refused surgery due to personal reasons.

The interaction between seizures and autonomic nervous system is very complex. Abnormal neuronal electrical activity arising from autonomic centers can result in autonomic symptoms anytime in the peri-ictal period. Episodic autonomic dysfunction is sometimes difficult to diagnose as seizures. Piloerection which may be accompanied by cold shiver is a very uncommon ictal sign of visceral epilepsies and may be isolated or occur in combination with other autonomic signs or complex partial seizures.[1,2] About 106 cases of pilomotor seizures have been reported in literature.[1-13] Out of these, 31 had isolated pilomotor phenomenon without any other aura or dialeptic features. The majority of patients reported in literature were noted to have ictal onset over the temporal region.[1,2,6,7,10] Piloerection is mostly unilateral or initially unilateral with later contralateral spread, and at times it can be bilateral at onset. Although majority had left temporal localization, one-third (35 cases) had right temporal localization.[1-13] On the other hand, bilateral piloerection had no definite lateralizing value.[1] Varied etiology of pilomotor seizures includes tumors, encephalitis, stroke, neurodegenerative disease, hippocampal sclerosis, or autoimmune process. Although hippocampal sclerosis is a more frequent cause,[1,2,7] a few recent reports have recorded pilomotor seizures in autoimmune nonparaneoplastic limbic encephalitis especially in relation to voltage-gated potassium channel antibodies.[6,10]

In temporal lobe epilepsy, piloerection, and other autonomic phenomena are proposed to reflect seizure spread to the insula.[1] An interesting observation is the high frequency of ictal pilomotor events with multiple daily to weekly events up to 20–30/h.[1,3,5-13] The exact pathophysiology of this high frequency of neuronal hypersynchronization is unclear. The presence of dysplastic cells, inflammation or involvement of limbic circuit including amygdala, mesial temporal and probably orbitofrontal structures may trigger increased seizure frequency due to high epileptogenicity index. We postulate that a reverberating circuit between limbic regions and centers of autonomic control may lead to a vicious cycle wherein pilomotor seizures and panic/anxiety or behavioral changes may be perpetuating each other. Further research using stereoelectroencephalography looking at the epileptogenic network involved in pilomotor seizures may answer this question. The peculiar features of ictal piloerection may masquerade as other psychiatric or cardiac disorders [Table 1]. The conglomerate of symptoms of gooseflesh, tachycardia, palpitations and sweating in themselves can easily be confused as neurovegetative dysfunction due to possible nonepileptic events.[5-8,10,11] As a consequence of the ictal phenomena and high frequency of recurrent events, personality and behavioral traits may develop simultaneously, further confusing the diagnosis. Eleven patients have been reported in literature to have initial misdiagnosis, 5 as due to psychiatric cause,[5,6]

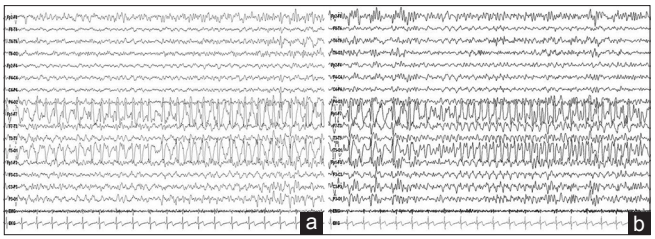

Figure 1: (a and b) Ictal electroencephalography shows rhythmic spike wave discharges followed by 5–6 Hz theta activity over left temporal region

one as cardiac and one as migraine. This leads to a significant delay in diagnosis with patients undergoing elaborate investigative tests and treatment for other disorders. The label of a psychiatric disorder may negatively impact the quality of life of the patient. Therefore, it is of paramount importance to suspect an epileptic etiology when encountering stereotyped, short-lasting, episodic events, howsoever bizarre, and uncommon in symptomatology.

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Conflicts of interest

There are no conflicts of interest.

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