

Two Siblings with Hot Water Epilepsy

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ABSTRACT Hot water epilepsy is a rare form of reflex epilepsies which occurs while bathing. Pathophysiology of this clinical entity is complex and unknown. It is supposed that a defect in the thermoregulatory system leads to the seizures due to rapid rise of body temperature. In this report, two siblings, one of whom is 35 year old male and the other 39 year old female, who have complex partial seizures that are triggered by contact with hot water during bathing are presented. The aim was to review the relevant literature in view of these two cases diagnosed with hot water epilepsy.

Keywords: Epilepsy, reflex; electroencephalography; epilepsy, complex partial

Reflex epilepsies are seizures precipitated by specific sensory stimuli and account for 6% of all epilepsies. Hot water epilepsy (HWE) is a rare form of reflex epilepsies triggered by specific thermal cutaneous stimulus. Different seizure types, both partial and generalized, may occur in HWE, suggesting common afferent pathway involvement. Familial clustering indicates that genetic factors may play a role in the development of the disease.¹ In previous studies, genetic locus of the disease has been defined on 4th, 9th and 10th chromosome.^{2,3} The diagnosis of the disorder is usually based on clinical history. Interictal electroencephalography (EEG) and neuroimaging studies are usually normal.

CASE REPORT

Thirty five years old male patient presented with déjà vu like symptoms which occur when taking a bath with hot water and feels himself as re-experiencing beautiful childhood memories. These symptoms were triggered by particularly contact of hot water to his face, lasted in approximately 1 minute and accompanied by short term loss of consciousness. His bathing habit was pouring hot water from a bucket with a mug over his head and he did not describe a seizure while washing his face without bathing. Patient stated that he felt a pleasure during seizures. Self induction and postictal confusion were not described. He described exactly the same seizures twice spontaneously within the last one year without any relation with bathing. His elder sister also experience similar bathing seizures. Thirty nine year old female patient described complex partial seizures without an aura during

bathing which started in childhood, triggered by hot water. During seizures, loss of consciousness, bizarre behaviours like shouting, irrelevant speech and symptoms consistent with psychomotor agitation occurred. Patient described short term postictal confusion. These seizures were stereotyped and lasted in 1-2 minutes. Her bathing habit was also the same as his younger brother, by pouring hot water over the head. She experienced no spontaneous seizures. Both of the patients stated that the longer they take bath the more the risk of seizure increases. Both patients', EEG and magnetic resonance imaging (MRI) were normal. As spontaneous seizures occurred in the male patient, carbamazepin treatment was initiated. In the female patient, antiepileptic treatment was not initiated. Lowering the heat of bathing water, shortening the bathing time and taking shower instead of pouring water over the head were recommended. Both patients are still being followed without any seizures. Patients-informed consent for publication was obtained.

DISCUSSION

Reflex epilepsies account for 5% of the seizures occurring in adults and 10% of those occurring in childhood.⁴ HWE is a rare form of reflex epilepsy and occurs most commonly in India and Turkey owing to genetic susceptibility and bathing habits.^{1,5-7} It accounts for 0.6% of all epilepsies, while in India it is around 4.4%.^{8,9} Its onset is usually in childhood and occurs more frequently in males than females.^{1,5-7,10} Seizures mostly arise due to pouring of hot water over the head whereas in 10% of patients immersion of body in hot water may trigger the seizures.¹¹ However, cases triggered by bath water at ambient temperature have also been reported.¹² In both of our cases, seizures occurred due to hot water contact in face. Of the seizures occurring in HWE, 67% are complex partial and 33% are generalized tonic clonic.^{6,8} It has been reported that in 1/3 of HWE cases, spontaneous seizures may also occur within 1-3 years.⁹ In our patients, consistent with the literature, complex partial type seizures were observed and spontaneous seizures were seen twice in the male patient without any relation with hot water.

The pathophysiology of HWE remains unclear. Satishchandra et al. suggested that HWE patients have genetically aberrant thermoregulatory systems and hence become susceptible to sudden increases in heat. They also reported that the return of body temperature to baseline levels after taking bath takes longer time than normal individuals.¹³ Although it was proposed that mean temperature of hot water which may trigger epileptic seizures is 41.4 C, water at optimal or normal heat may be high enough to trigger paroxysmal seizures.^{12,14} A complex combination of tactile and heat stimulants are required in order that such seizures can take place. In a study, although seizures were triggered in laboratory environment by pouring hot water over the head of patients, seizures were not stimulated by placing hot towels on head, in sauna environment or being exposed to hot water. Therefore, it was suggested that triggering factors for this rare form of epilepsy are quite complex and that they may appear with the combination of factors such as contact with hot water, water temperature and involvement of specific cortical region by the stimulus.¹⁰

HWE may be sporadic or familial. In studies carried out in India, the rate of family history ranges between 7-18%.¹¹ In two studies from Turkey, family history was found at the rates of 10% and 55.6% respectively.^{8,15} In familial type, genetic transmission is not clearly known. However, recently a genetic locus of the disease transmitted dominantly at 4th, 9th and 10th chromosomes was defined.^{2,3} These findings indicates genetic heterogeneity for the disorder.

In HWE, pleasure may be felt during seizures. It was found that in 42.8% of the patients describe a pleasant feeling during seizures and that about 1/3 of the patients self induce their seizures.⁸ In recent case, male patient described feeling pleasure during seizures but did not mention self induction.

Interictal scalp EEG is usually normal, but 15-20% might show diffuse abnormalities.¹¹ Obtaining ictal EEG is technically quite difficult. When it is possible, paroxysmal discharges characterized by

focal epileptic activity and secondary generalization in temporal and frontal regions might be seen.⁸ In HWE, neuroimaging findings are usually normal. In some case reports, structural lesions such as occipital cortical dysplasia, mesial temporal sclerosis, temporal arachnoid cyst, hippocampal atrophy and partial cortical dysplasia have been described.^{9,16} Ictal single photon emission tomography (SPECT) showed hyperperfusion of the temporal lobes in some of the patients with HWE.¹⁷ In neither of our cases, abnormality was found in interictal EEG and brain magnetic resonance imaging studies. Ictal EEG could not be obtained.

In HWE spontaneous remission may also develop. In the first step of treatment, precautions such as lowering the heat of bath water and not pouring hot water over the head are relevant. In conditions when these precautions fail to prevent seizures or spontaneous ones occur, antiepileptic treatment may be required. A prospective long term study has confirmed, intermittent prophylaxis prior to head water bath with oral clobazam for the management of pure HWE.¹⁸ Our male patient underwent carbamazepin treatment since spontaneous seizures were also present and these seizures were controlled by treatment. In our female patient, decreasing the heat of bathing water sufficed and antiepileptic treatment was not initiated as she did not experience spontaneous seizures.

As HWE occurs due to exposure to hot water during bathing and seizures are mostly of complex partial type, they are not easily recognized by patients and their relatives. Therefore, they usually do not seek medical help. However, definite diagnosis is especially crucial that, these seizures can be prevented by a few protective measures. Genetic factors also play part in the pathogenesis of the disease, which makes it important to question family members with respect to similar symptoms.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Aşlı Ece Çilliler; **Design:** Aşlı Ece Çilliler; **Control/Supervision:** Hayat Güven; **Data Collection and/or Processing:** Aşlı Ece Çilliler; **Analysis and/or Interpretation:** Aşlı Ece Çilliler; **Literature Review:** Aşlı Ece Çilliler; **Writing the Article:** Aşlı Ece Çilliler; **Critical Review:** Selçuk Çoçmoğlu.

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