

HOT WATER EPILEPSY (HWE) FAMILIAL AND NONFAMILIAL CASE REPORTS

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ABSTRACT

Reflex epilepsy is a form of epilepsy precipitated by a specific stimulus and events. Seizure precipitation by hot water during bathing is named as "hot water" or "bathing epilepsy" which is a rare and unique form of reflex epilepsy.

It is especially common in Southern India and of higher frequency in Muslims among them. There are also isolated and rare case reports from Australia, Japan, Canada and the United States. A small series of patients with hot water epilepsy (HWE) was reported from Turkey. The cases with HWE may be sporadic or familial.

Our aim is to outline the clinical and electroencephalographic (EEG) features of four patients with hot water epilepsy, seen at our clinic. One of our patients has a family history of epilepsy. The second and third generation of family members had seizures during bathing.

It is important to ask patients with HWE about bathing habits (pouring hot water over the head from a bowl), the temperature and amount of bathing water, the duration of bathing, any additional spontaneous seizures and family history. Sometimes this is enough to eliminate precipitating factors for treatment.

Keywords: Reflex epilepsy, Hot water, Pathophysiology, Treatment

SICAK SU EPİLEPSİSİ AİLESEL VE SPORADİK OLGU SUNUMLARI

ÖZET

Refleks epilepsi iç ve dış uyaranlar veya etkenlerle oluşan bir epilepsi türüdür. Banyo sırasında sıcak su ile tetiklenen nöbetler "sıcak su" veya "banyo epilepsisi" diye adlandırılır ki refleks epilepsinin nadir rastlanan bir formudur.

Özellikle Güney Hindistan bölgesinde yaygın olup, Müslümanlar arasında sıklığı daha fazladır. Bunun dışında Avustralya, Japonya, Kanada ve Amerika'da da izole, nadir olgular bildirilmektedir. Ülkemizede de sıcak su epilepsine ilişkin küçük bir seri mevcuttur. Sıcak su epilepsisi sporadik veya ailesel özellik gösterebilir.

Bu çalışmada polikliniğimizde takip edilen sıcak su epilepsili 4 olgunun klinik ve elektroensefalografik (EEG) özelliklerini ortaya koymak amaçlanmıştır. Hastalarımızdan birinde aile hikayesi de bulunmaktadır. Hastanın 2. ve 3. kuşak akrabalarında banyo yaparken nöbet geçirme söz konusudur.

Sıcak su epilepsisi olan hastalara banyo yapma alışkanlıkları (sıcak suyu bir tas ile başlarından aşağı dökme gibi), suyun sıcaklığı, banyoda kalma süresi, sıcak su ile tetiklenen nöbetlerin dışında spontan nöbetlerin olup olmadığı, aile hikayesinin bulunup bulunmadığının sorulması önemlidir. Bazen tedavide sadece tetikleyen faktörlerin ortadan kaldırılması yeterli olmaktadır.

Anahtar Kelimeler: Refleks epilepsi, Sıcak su, Patofizyoloji, Tedavi

Marmara Medical Journal 2005;18(3);131-134



INTRODUCTION

Reflex epilepsy is a form of epilepsy provoked by a specific stimulus or event. It is only 6% of all epilepsies^{1,2}.

Reflex epilepsy is a collection of different seizure types and their specific provoking stimuli². Photosensitive epilepsy (induced by light stimuli) is the most common type². Other than this, seizures induced by thinking, reading, eating, shocks, music or hot water are also classified as reflex epilepsy^{1,2}.

Seizure precipitation by hot water during bathing is named "hot water" or "bathing epilepsy" which is a rare form of reflex epilepsy. I.M.Allen, in 1945, from New Zealand first described a 10-year-old boy who had seizure during bathing. Other case reports followed from Japan, Canada, Australia, and the United States. The largest series, 279 patients, have been reported from Southern India. Although the percentage of patients with HWE was 0.6% among all patients with epilepsy, in Southern India HWE patients were reported as 3.6-3.9%³⁻⁵.

It is possible to provoke the seizure by pouring hot water over the head in the bath. The temperature of water used ranges between 40-50°C and the room temperature is 25-30°C^{4,5}. The majority of patients are male and children. A positive history of epilepsy among family members has been reported in 7-20% of cases. Spontaneous non-reflex epilepsy is reported in 25% of patients^{4,6}. The pattern of epileptic seizure is generally complex partial form but generalized tonicclonic seizure (GTCS) is also reported³.

CASE REPORTS

Case Report 1

A 23-year-old male patient reported to have a GTCS when he took a bath with hot water especially when he spent a long time in the bath.

When the duration of bathing was longer, I have a seizure consequently. But when I am taking a hot bath in a short time or with warm water, I do not have any problem. And also, when I have a shower without pouring water directly on my head, I have no seizure, either"......

Some of his second and third generation of family members have seizures during bathing.

His neurological examination, MRI and interictal EEG were all normal and the seizures were controlled when the provocating factor was eliminated.

Case Report 2

An 18-year-old female patient has seizures while having a bath.

"While my mother was bathing me with hot water, I was getting a pleasant feeling when I looked at one point on our bathroom's wall. And then I had been turning around myself unpurposely. After 7 years old, I realized that when I did not look at a point during bathing I did not have seizures. But when I was 14 years old, looking at a point during bathing gave me a pleasure. So I kept looking at one point on the wall to live this experience again and again".....

This patient's MRI, neurological examination and interictal EEG were also normal.

Case Report 3

An 18 year-old male patient presented with history of seizure while taking a bath with hot water since the age of three. He used to become motionless with activity arrest. He explained that the precipitating factors were the water's temperature and pouring it directly over his head.

There was no history of seizures during any febrile illness, head injury or similar complaints in the family.



The patient's neurological exam was normal but he had not neuroimaging. His interictal EEG showed mild and generalized disorganized background.

Case Report 4

A 19 year-old female patient admitted to our hospital for the first episode of seizure while she was taking a bath.

"After the dinner I was taking a shower, but the water was hot and the time I spent was about 10 minutes. I felt something getting wrong and I decided to open the window to breath comfortably. And I did not remember anything after that. It was the first episode I experienced. My mother found me in the bathroom in a tonic-clonic movement with a ' cyanotic appearance'.

The patient's neurological examination and MRI were normal.

Her sleep deprivation EEG showed epileptiform activity and she was advised to take bath with warm water. Valproic acid (VA) treatment was begun for the control of her seizure.

DISCUSSION

Hot water epilepsy refers to a specific type of reflex epilepsy. HWE is considered to be a geographically specific epileptic syndrome since it mainly occurs in Indian'. HWE is mostly seen in the first decade of life. It is more common in children, with cases more frequent among male than female patients $(70\%)^4$. The seizure could occur either at the beginning or at the end of the bath and last about 1 to 3 minutes. The pattern of epileptic seizure, which is seen in HWE, consists of 67% complex partial (CPS) and 33% generalize tonic clonic seizure (GTCS). In a different study CPS was reported in 40%, GTCS in $60\%^{3,4}$. Gururaj G, et al., noticed that the frequency of seizure occurrence was more than 1-4 attacks/month in 89% of cases⁸. Symptoms onset included a dazed look, a sense of fear, irrelevant speech, and visual and auditory hallucinations with complex automatism⁴.

Interictal EEG is generally normal but in 20-25% of cases show diffuse abnormalities. The factors that provoke seizures are the temperature and the amount of water, the body surface of pouring water, the type and the duration of bathing³.

In our cases the main precipitating factors for seizures were bathing with hot water and pouring water over the head. It seems that traditional factors are important here, because Turkish people have a bath by sitting and pouring water with a bowl³.

In some patients, seizures are only activated by hot water, but spontaneous non-reflex epilepsy is also reported in different series from 16-38% to 62% of patients^{3,4,6}. Twenty-five percent of cases develop nonreflex epilepsy within 1-3 years⁹.

Five to ten percent of patients had seizures even during a body bath, when water is not poured overhead. Eleven to twenty-seven percent of patients had febrile convulsions prior to the development of reflex epilepsy. A positive history of epilepsy among the family members had been reported in 7-20% of cases^{4,6}.

The mechanism of reflex epilepsy is not clear. A defect of thermoregulation center in the hypothalamus might be the underlying mechanism of reflex epilepsy. When a hotwater epileptic patient takes a bath, his body temperature increases 2-3°F in the short span of 2 minutes and returns to the baseline after 10-12 minutes when the bathing is completed. On the other hand, when normal healthy volunteers have a bath, the body temperature raises 0.5 to 0.6°F and returns to baseline immediately at the end of the bath. So, HWE patients probably have an aberrant thermoregulatory system and are extremely sensitive to the rapid rise in temperature. This aberrant thermoregulation system seems to be genetically determined⁴.

The cases with HWE may be sporadic or familial. In familial type, the transmission of inheritance is not known. But it may be



genetically determined or only related to traditional bathing habits^{3,4}. In our clinic, we have encountered one patient with a positive family history of HWE .

Patients with HWE reported feeling pleasure during their seizure and nearly 30% of them triggered their seizure by themselves¹⁻³. Case 2 has also mentioned this feeling of pleasure. HWE is generally known as benign and selflimited, but sometimes antiepileptic drugs are necessary for controlling seizures. Avoidance from hot water may be sufficient to be seizure free. If it is necessary to take a medication,VA and carbamazepine (CBZ) might be preferred. Rectal diazepam (DZP) may also be given 30 minutes before bathing^{3,6}.

Some of our patients are still not taking any medications. They are taking their bath in a shorter time and with warm water.

HWE is a benign form of reflex epilepsy. It is important to take clinical history in detail for diagnosis and control of seizures, without medications if possible.

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